J Opthalmol Res 2021; 4 (3): 254-257 *DOI: 10.26502/fjor.2644-00240043*

Research Article

Visual Evoked Potential Findings of Bardet - Biedl Syndrome

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Received: 05 August 2021; **Accepted:** 11 August 2021; **Published:** 13 August 2021

Citation: Fatemeh Ojani, Seyed Mohammad Masoud Shushtarian, Ahmad Shojaei, Jalil Naghib. Visual Evoked Potential Findings of Bardet - Biedl Syndrome. Journal of Ophthalmology and Research 4 (2021): 254-257.

Abstract

Purpose: Bardet – Biedl syndrome (BBS) is a complex disorder that affects many parts of the body including visual pathway of visual system. The purpose of present work is to prove the adverse effects of BBS on visual pathway using visual evoked potentials.

Patients and Methods: Ten patients, 6 males and 4 females with age range of 12 - 20 years were selected. The latency and amplitude of visual evoked potential, P100 peak was recorded for these patients and 10 age and sex matched controls with healthy visual system mainly visual pathway.

Results: The mean age was 15.4 ± 3.06 and 15 ± 2.78 in case and control group respectively. The mean visual acuity in case 0.49 ± 0.14 (LogMAR) and 0 ± 0 (LogMAR) in control group which was statistically significant in two groups which is the characteristics of BBS patients. These was statistically significant higher latency and lower amplitude of visual evoked potential, P100 peak in case group compared control groups ($P < 0.001$)

Conclusion: BBS have adverse effect on visual system mainly visual pathway causing increased latency and decreased amplitude of VEP, P100 peak measured using visual evoked potential.

Keywords: Bardet - Biedl Syndrome; Visual pathway; Visual evoked potential

1. Introduction

Bardet Biedl syndrome (BBS) is a rare genetic disorder with highly variable symptoms which may include retinal degeneration. obesity, reduced kidney function, Polydactyly among many other features. Visual system is among, the organs which may be affected in BBS Patients. Visual acuity fall, strabismus, Cataracts and glaucoma are among the eye diseases observed in some individual. Visual acuity disturbances is more common in BBS Patients. In this regard Adriana Berezovsky on 2012 worked on visual acuity of a group of 23 Patients (15 males) with ages ranging from 6-36 years (mean 15.8 ± 6.4; median=14.07) was assessed. Visual acuity in the better- seeing eye was 20/40 or better in 5 Patients (21.7%). 20/50 - 20/150 in 13 (56.5%) patients. 20/200 - 20/400 in 2 (8.7%) patients and worse than 20/400 in one (4.3%) patients. The mean visual acuity in the better seeing eye was 0.7 ± 0.6 logMar (20/100. Snellen equivalent) [1]. It is observed that most patients with BBS will experience the loss of a particular population of cells in the eyes specifically in the retina. This produces gradual loss of rod and cone cells in retina. Symptoms associated with rod-cone dystrophy may not become apparent until 7 or 8 years of age when children begin to complain of an inability to see in dimly lit environment. Affected individual often first lose peripheral vision and this situation may Progress to central vision too; In some patients the degeneration of retina may follow a characteristics Course, referred to as retinitis Pigmentosa (RP). To screen the retina, especially when the rod-cone cells are degenerated electroretinography is a suitable electrophysiological technique. In this regard, Naser M, and her colleagues 2020 worked on fifty RP patients. They were undergone ERG examination. The result showed pathological changes in amplitude and latency of ERG a and b peaks of patients in comparison to control group and they concluded that ERG is a suitable technique to look for pathological changes observed in retina of RP Patients [2]. Another supporting study was reported on 2014 by Tahmasebi S, and her research team. They also reported the usefulness of ERG as a technique to diagnose the retinal status of RP patients [3]. Recently on 2021, an extensive work was carried on visual pathway of RP Patients using several techniques including visual evoked potentials (VEP) and they concluded that visual pathway of these patients are impaired [4]. So, an effort was done to search for possible visual pathway changes of BBS Patients using VEP technique.

2. Patients and Methods

In this case control study ten (6 male + 4 female) Patients (20 eyes) with BBS were selected as the case group. They were in age range of 12-20 years. The Visual evoked potential test with checkerboard stimulation method was performed to evaluate the visual pathways of Patients. Latency (msec) and amplitude (µV) of VEP, P100 Peak was measured for all participants using Mangoni machine. In summary three electrodes were used to connect the machine to the patients. Active, reference and ground electrodes were attached to occipital, Vertex and forehead of patient respectively. The same procedure was repeated for 10 age and sex matched healthy individual (20 eyes) as the control group. The results obtained in two groups were compared for Probable differences between the two groups.

3. Results

Variable	Number of participants	Groups (Mean ± SD)		P value
		Case	Control	
Age	20	15.4 ± 3.06	15 ± 2.78	0.796**
sex	20	-	-	0.653*
VA LogMar	20	0.49 ± 0.14	0 ± 0	0.000**

*Based on chi-square

**Based on Mann-Whitney Test

Table 1: Demographic findings in the case and control groups and there was no statistically significant difference between the two groups regarding the sex (P = 0.653) and age (P = 0.796) where as there is statistically significant difference between two groups as far as visual acuity is concerned (P <0.001).

Variable	Number of participants	Groups (Mean ± SD)		P value*
		Case	Control	
latency (msec)	20	116 ± 3.88	98.8 ± 2.85	0
Amplitude (µv)	20	2.2 ± 1.03	8.4 ± 1.83	0

* Based on Mann-Whitney Test

Table 2: Measurements of mean latency and amplitude of VEP, P100 peak in the case and control groups.

Table 2 shows the measurement results for latency and amplitude of VEP, P100 peak in the case and control groups. There was statistically significant higher latency and amplitude of the visual evoked potential, P100 peak in the case group compared to the control group (P < 0.001).

4. Discussion

A research was set to look for visual pathway status of BBS patients. The two groups i.e., control and case were similar as far as demographical aspects were concerned i.e., age and sex. It was observed that visual acuity was quite low in patients' group, which is the characteristics of these patients and was reported by other [1]. VEP results indicate prolonged VEP, P100 Peak in BBS Patients in comparison to healthy group whereas the amplitude was quite low in case group rather than control group. It is a fact that VEP is the integration of visual pathway [5, 6] so the changes in the VEP can be the reason for pathological changes in visual pathway of BBS Patients. The result of present work may be supported by following research work. Harw A Tabl worked on 3 female patients (9, 10 & 16 years old) with BBS and were complaining of Progressive visual loss. He tested different electrophysiological technique including Pattern VEP on the patients. All the tests showed abnormal results & specifically pattern VEP showed prolonged latency & reduced amplitude of VEP P100 wave, which is in accordance to result of present work [7]. Another work in this connection was done by Xiaohong Meng and his coworkers on 2021. They worked on 12 BBS patients. All patients had typical Phenotypes of retinitis pigmentosa with unrecordable or severely damaged cone and rod responses on full field flash electroretinography (ffERG). Most of the patients showed unremarkable reaction in Pattern visual evoked potential (PVEP) and multifocal electroretinography (mfERG) while their flash visual evoked potential (FVEP) indicated display residual visual function and they demonstrate the usefulness of a combination of ffERG & FVEP assessments of visual function in the advanced stage of retinopathy in BBS [8]. This

reference once again is a clue for the reliability of present work. The only difference is that they could not find remarkable reaction in PVEP where as we could find it. The reason is nothing but the severity of damage in retina, where as in present research the extent of damage was not to that stage not to record PVEP. One more reference in BBS patients is a review article deals with genetics, molecular pathophysiology and diseases management in these patients. The authors stated that visual impairment in BBS has been consistently early in onset; between 8 and 9 years and 98% of the patients suffered complete loss of vision by the third decade. They further reported that ERG and VEP are often normal up to 5 years of age [9]. As far as VEP is concerned in present work, the age range of patients were between 12 to 25 years so the changes in VEP's are most probable.

5. Conclusions

Bardet - Bidle syndrome (BBS) might have adverse effect on visual system mainly visual pathway, causing increased latency and decreased amplitude of VEP, P100 peak measured using visual evoked potential.

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